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Solitary Fibrous Tumor Arising from Stomach: CT Findings

Sung Hee Park,¹ Myeong-Jin Kim,^{1,2} Jieun Kwon,³ Jong-pil Park,³ Mi-Suk Park,¹ Joon Seok Lim,¹
Joo Hee Kim,¹ and Ki Whang Kim¹

¹Department of Diagnostic Radiology, Severance Hospital and Research Institute of Radiological Science, ²Institute of Gastroenterology and Brain Korea 21 Project for Medical Science, ³Department of Pathology, Severance Hospital and Research Institute of Pathological Science, Yonsei University College of Medicine, Seoul, Korea.

Solitary fibrous tumors are spindle-cell neoplasms that usually develop in the pleura and peritoneum, and rarely arise in the stomach. To our knowledge, there is only one case reporting a solitary fibrous tumor arising from stomach in the English literature. Here we report the case of a 26-year-old man with a large solitary fibrous tumor arising from the stomach which involved the submucosa and muscular layer and resembled a gastrointestinal stromal tumor in the stomach, based on what was seen during abdominal computed tomography. A solitary fibrous tumor arising from the stomach, although rare, could be considered as a diagnostic possibility for gastric submucosal tumors.

Key Words: Solitary fibrous tumor, stomach, CT findings

INTRODUCTION

Solitary fibrous tumors are rare neoplasms of mesenchymal origin that account for less than 2% of all soft tissue tumors and have classically been described in the thorax as arising from the pleura.¹⁻³ However, they have been found in extra-serosal sites and should be considered in the differential diagnosis of any spindle cell lesion, including those in the gastrointestinal tract.³ In the pleural site, solitary fibrous tumors have unique gross and microscopic appearances and can be easily recognized. When they arise at other non-pleural sites, differentiation from other soft tissue

tumors can be difficult.⁴

To our knowledge, only one case of a solitary fibrous tumor arising from the stomach has been reported,⁵ but its radiological findings have not been well described.

We performed computed tomography (CT) and here we report the findings from a solitary fibrous tumor arising from the gastric wall that mimicked a gastrointestinal stromal tumor.

CASE REPORT

A 26-year-old man presented with melena and underwent upper gastrointestinal endoscopy and abdominal CT.

Endoscopy showed a large submucosal tumor in the gastric body with concomitant bleeding from a large central ulceration containing fluid and residual contrast material from a previous barium study (Fig. 1A). Endoscopic ultrasound similarly showed a large submucosal mass (Fig. 1B). A first endoscopic biopsy result was non-specific and showed acute and chronic inflammation with ulceration. The second biopsy revealed ulceration and tissue granulation with foreign body giant cells suggestive of a barium granuloma.

An abdominal CT scan demonstrated about 5.5 × 3.2 cm sized, well-defined and large exophytically growing mass arising from the posterior aspect of the lesser curvature side in the gastric body. The mass showed a large ulceration at the gastric luminal side and a cavity in the central portion communicating with the gastric lumen.

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Reprint address: requests to Dr. Myeong-Jin Kim, Department of Diagnostic Radiology and Research Institute of Radiological Science, Institute of gastroenterology, Yonsei University College of Medicine, 250 Seongsanno, Seodaemun-gu, Seoul 120-752, Korea. Tel: 82-2-2228-7400, Fax: 82-2-393-3035, E-mail: kinnex@yuhs.ac

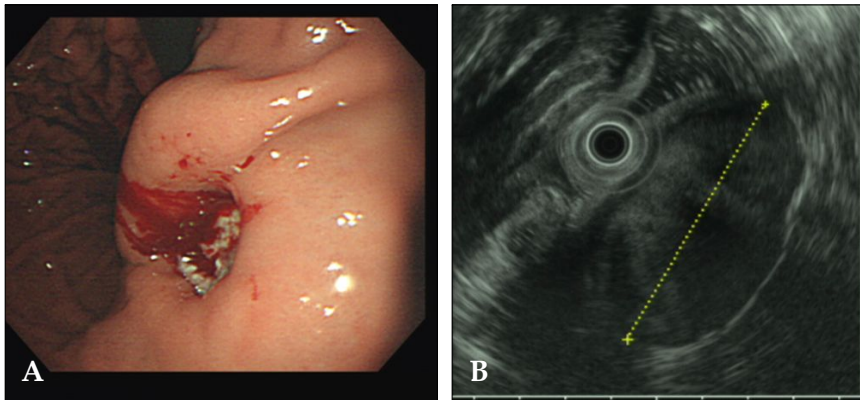


Fig. 1. (A) Endoscopy showed a large submucosal tumor in the gastric body with bleeding from a large central ulceration that contained fluid and residual contrast material from a previous barium study. (B) Endoscopic ultrasound (EUS) also showed a large submucosal mass in the gastric body portion.

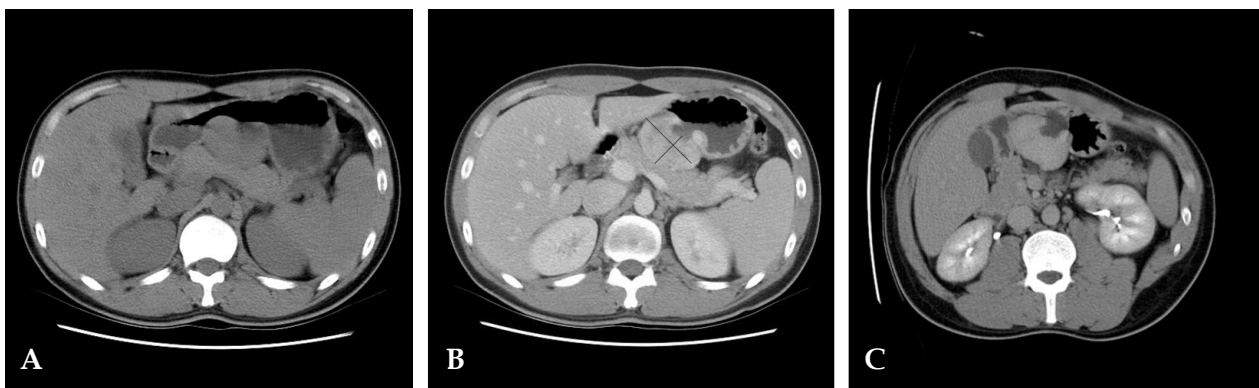


Fig. 2. Abdominal computed tomography (CT) demonstrated about 5.5×3.2 cm sized, well defined large mass arising from the posterior aspect of the lesser curvature side of the gastric body. (A) The mass showed relative hypoattenuation on the precontrast images. (B) In the portal phase, the mass showed intense heterogeneous enhancement. Ulceration in the luminal side and low attenuation in the central cavity were seen within the mass. (C) In the equilibrium phase, the mass showed prolonged enhancement and the bulk of the tumor was seen in an extragastric location.

The bulk of the tumor was seen in an extragastric location with extensions into the gastrohepatic ligament and lesser sac. There was no evidence of direct invasion of the adjacent organs or peritoneal spread. No metastatic lesions or lymphadenopathy was seen in the abdomen or pelvis. The mass showed relative hypoattenuation on the precontrast images (Fig. 2A) and increased heterogeneous enhancement in the portal phase (Fig. 2B) and prolonged enhancement in the equilibrium phase images (Fig. 2C). The radiologic findings were distinct from those of epithelial tumors and the possibility of a malignant gastrointestinal stromal tumor of the stomach was considered on CT. The mass was removed under general anesthesia by laparoscopic wedge resection of the stomach. During laparotomy there was a round and well demarcated mass arising from the lesser

curvature side of the gastric body, and there were prominent vessels around the mass with no detected ascites and no adhesion to the adjacent structures. On gross pathology, the specimen consisted of a portion of the stomach measuring 8.5×5 cm with an attached ovoid and lobulated rubbery to firm solid mass measuring $5.4 \times 5.2 \times 4$ cm. The mucosa of the stomach was retracted by underlying the attached mass. Sectioning of the specimen revealed a well demarcated and relatively homogeneously yellow colored mass involving the submucosa and muscular layer (Fig. 3).

Microscopic examination indicated that the mass was composed of a patternless proliferation of bland spindle cells with abundant perivascular and intercellular mature collagen formation (Fig. 4A and B). There were few mitotic cells and no

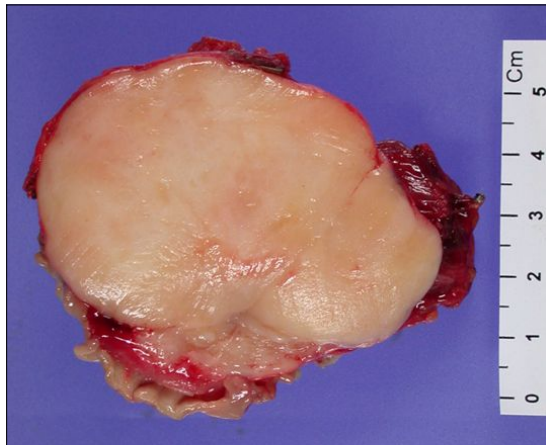


Fig. 3. On gross pathology, the specimen was an ovoid, lobulated solid mass, measuring $5.4 \times 5.2 \times 4$ cm, which was attached to the stomach as a portion of the stomach. On sectioning of the specimen, it was a well demarcated and relatively homogeneously yellowish mass involving submucosa and muscular layer.

nuclear atypia observed in 10 high-power fields, nor any tumor necrosis, suggesting high grade malignant degeneration.

Immunohistochemistry showed that the neoplastic cells were strongly and diffusely positive for CD34 and negative for c-kit (CD117) (Fig. 4C and D), and they were also positive for S-100 and negative for both smooth muscle actin and desmin. Because solitary fibrous tumor rarely develop in the stomach, kit negative GIST was considered as its alternative and PDGFRA gene analysis was performed. However, there was no mutation in the result of PDGFRA gene analysis. Although S-100 positive finding can be seen in neurogenic tumors such as schwannoma, there were no histologic findings suggestive of neurogenic tumor such as nuclear palisading, whorling of the cells and Verocay bodies. Moreover, CD34

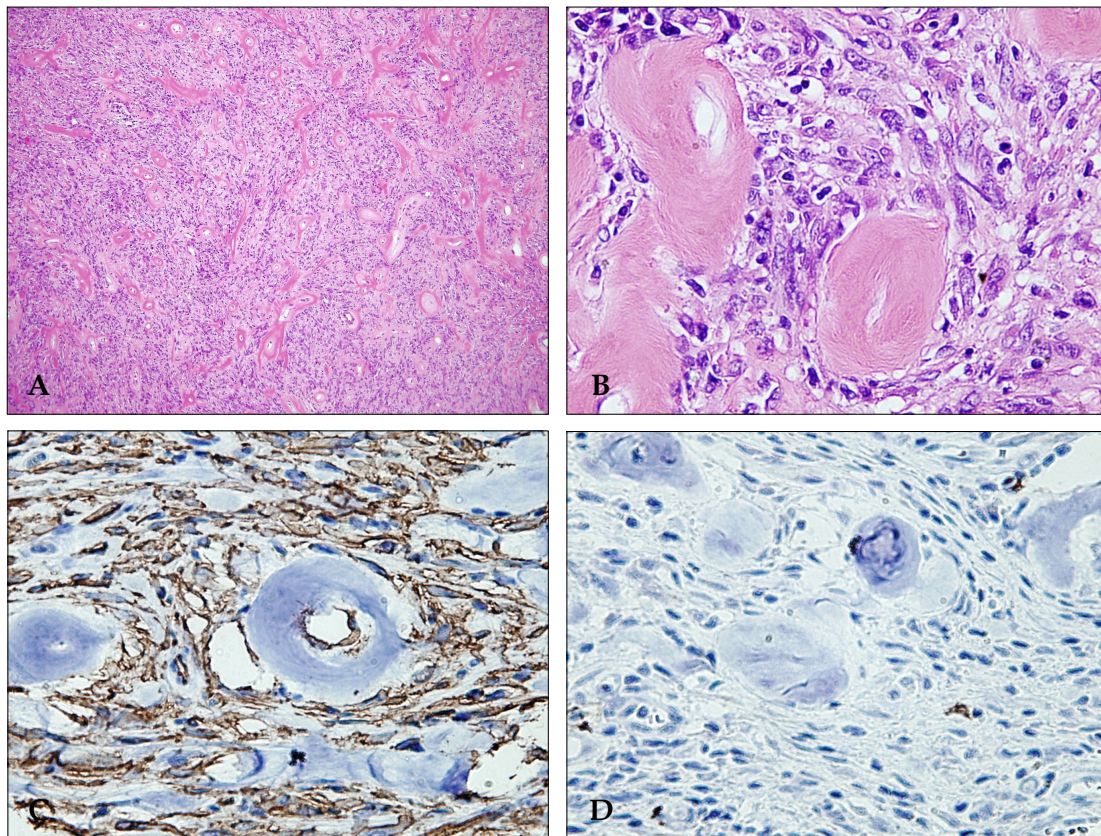


Fig. 4. Histologic findings by Hematoxylin and Eosin [H-E] stains (A, B) and immunohistochemical findings for CD34 and c-kit. (C, D). (A) Photomicrograph (original magnification, $\times 100$; H-E stain) shows that the patternless proliferation of spindle cells with abundant perivascular and intercellular mature collagen formation. Dilated, thick-walled vessels were common in the lesions. (B) High power photomicrograph (original magnification, $\times 400$; H-E stain) shows that non-atypical, round to spindle-shaped tumor cells have little cytoplasm with indistinct borders and dispersed chromatin within vesicular nuclei. There was rarely mitosis. (C, D) The tumor cells showed strong immunoreactivity characteristic of neoplastic cells for CD34 (immunoperoxidase, $\times 400$) (C) and the tumor was negative for c-kit ($\times 400$) (D).

test result was positive which are very exceptional in the case of neurogenic tumor, thus greatly diminishing the possibility of neurogenic tumor as its final diagnosis. Conclusively, all these findings are diagnostic for solitary fibrous tumors. The patient's melena resolved following removal of the tumor and the subsequent one year of follow up has revealed no evidence of recurrence or metastasis in this patient.

DISCUSSION

Solitary fibrous tumors most commonly affect the pleura but have been reported in other extrapleural locations.^{2,6}

The histological features of solitary fibrous tumors are very distinctive and remarkably consistent with typically conclusive immunohistochemistry and electron microscopy.⁷

Generally, these tumors are diagnosed pathologically by the presence of a collagenous matrix with arrays of spindle cells exhibiting diffuse CD34, bcl-2, and vimentin positivity, and S100, actin, and keratin negativity on immunohistochemical analysis.^{1,8} It is the unusual location rather than the microscopic appearance which causes difficulty in making diagnosis.⁷ The imaging characteristics and radiologic features of solitary fibrous tumors arising from unusual locations are not well established in the literature due to the rarity of these tumors.

Solitary fibrous tumors of the pleura typically demonstrate a well-defined, lobular, solitary nodule or mass. Small solitary fibrous tumors demonstrate homogeneous, well defined, non-invasive, lobular, soft tissue masses, which typically abut a pleural surface, and may form obtuse angles against the adjacent pleura. The majority of solitary fibrous tumors are histologically benign, but up to 20% of all solitary fibrous tumors may be malignant.^{1,9}

On CT scan, solitary fibrous tumors typically appear as well delineated, smooth, lobulated soft tissue masses that may occasionally contain scattered calcifications. Smaller tumors tend to demonstrate homogeneous enhancement, whereas central tubular or rounded low attenuation areas may be seen in larger lesions, representing cystic

or necrotic changes.^{7,8}

In the present case, the abdominal CT scan demonstrated a well-defined submucosal tumor arising from the gastric body. Radiological diagnosis indicated a gastrointestinal stromal tumor (GIST) arising from the stomach, because GIST is the most common mesenchymal neoplasm of the gastrointestinal tract and has many features similar to those present in our case. The stomach is the most common location for GIST, in which GIST makes up 2-3% of all gastric tumors.¹⁰ During CT, GISTs may have a variable appearance and attenuation based on size; small tumors tend to appear homogeneous and larger ones frequently show central low attenuation due to necrosis or hemorrhage.¹¹ When they are large (> 5 cm), the tumors often appear exophytic and may contain necrosis and calcification.^{12,13} More than 70% of gastric GISTs were located in the body.¹⁰ Associated adenopathy is uncommon.¹⁴

Other differential diagnosis possibilities for submucosal tumors include fibromatosis, inflammatory myofibroblastic tumors, schwannomas, leiomyomas, and leiomyosarcomas. Differential diagnosis of stomach mesenchymal tumors in the stomach using only CT imaging is difficult. Recently, advanced immunohistochemistry has allowed differentiation from other benign soft tissue tumors such as fibroblastic or neurogenic tumors.¹⁵ Immunoreactivity to CD34 is crucial to the diagnosis of a solitary fibrous tumor but CD34 is also expressed in other tumors and approximately 70% of GISTs coexpress CD34 with c-kit.^{16,17} In solitary fibrous tumors, neoplastic cells show negative reactivity for c-KIT. And in solitary fibrous tumor, focal and limited reactivity for S100 protein, cytokeratins and/or desmin has also occasionally been reported.^{18,19}

Extrapleural solitary fibrous tumors may present with symptoms related to the tumor site and size, or with systemic symptoms which include hypoglycemia, arthralgia, osteoarthropathy and clubbing.²⁰

In conclusion, we have reported a rare solitary fibrous tumor arising from the stomach. This lesion showed typical imaging features of a gastric submucosal tumor with similarities to GIST that are more common in stomach. Therefore, solitary fibrous tumors should be included in the list of

differential diagnoses possible for a gastric submucosal tumor.

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